Assessment and Management of Pes Cavus in Charcot–Marie–Tooth Disease

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The sequential approach to evaluating the cavus foot is integrated with a description and assessment of the various treatment options. Decision making in the treatment of these cases is complicated by the progressive neurologic condition that underlies many of these deformities. An effort is made to recommend the most appropriate surgical intervention based on the nature of the deformity and its rigidity. Although these principles apply to all cavus feet, the deformity in Charcot–Marie–Tooth disease is the most difficult to treat and the most prone to recurrence because of the progressive nature of the muscular imbalance causing it.

Pes cavus due to progressive neuromuscular disease is a challenging management problem. Two thirds of the patients who seek treatment for a painful high-arch foot will have an underlying neurologic problem, and half of these will have Charcot–Marie–Tooth (CMT) disease, an inherited degenerative disorder of the central and peripheral nervous systems that causes muscle atrophy and loss of proprioception.[1] CMT disease usually is an autosomal dominant trait, and a family history of thin legs and high arches frequently can be obtained. In more than 70% of patients with the autosomal dominant form, muscle atrophy is steadily progressive. Less often, the disease will arrest completely (23%) or be manifested intermittently (7%).[16] The X-linked recessive and autosomal recessive forms have an early onset (first decade and second decade, respectively) and are more rapidly progressive; severe involvement is usually present within a decade after appearance of the first manifestations.[11]

In CMT disease, the initial complaints are general foot weakness and unsteady gait. Specific foot complaints include pain under the metatarsal heads, claw toes, foot fatigue, and difficulty in fitting regular shoes. With autosomal dominant inheritance, 53% have mild pes cavus, and 22% have a severe cavus deformity.[11] Distal loss of proprioception and spinal ataxia are common. Patients with recessive forms of CMT disease usually are more disabled as a result of extensive involvement of proximal muscle groups. Scoliosis and marked loss of upper extremity function are frequent in patients with recessive inheritance.[11]

CMT disease should be suspected in patients with claw toes, high arches, thin legs, poor balance, and an unsteady gait. Evaluation by a neurologist is essential prior to considering orthopedic intervention. Electromyograms and nerve conduction studies showing slow sensory and motor conduction and selective muscle denervation, supported by appropriate physical findings and a positive family history, confirm the diagnosis. Spinal roentgenograms should be part of the diagnostic workup of all patients with progressive pes cavus.

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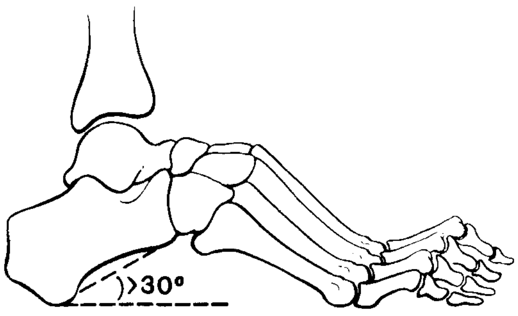


FIG. 1. Measurement of the calcaneal pitch angle.

PES CAVUS: DIFFERENTIAL DIAGNOSIS

Management of the cavus foot in CMT disease is complex. The progressive nature of the disease produces a wide spectrum of involvement at presentation. Formulation of a treatment plan starts with careful evaluation of the foot to distinguish fixed and flexible deformities. Important aspects of the deformity to be evaluated have been outlined by Japas,[9] Ibrahim,[7] and Samilson and Dillin.[15] Planning treatment, particularly surgical decision making, is facilitated by addressing the following questions: (1) Is the deformity unilateral or bilateral? (2) Is the cavus deformity primarily anterior or posterior? (3) Does the forefoot deformity involve the entire metatarsus or predominantly the first ray? (4) Is the hindfoot varus deformity fixed or flexible? (5) Are the claw toes correctable passively?

Unilateral involvement is rare in CMT disease. True unilateral pes cavus is often due to poliomyelitis or trauma. Deep posterior compartment syndrome[10] and crush injury of the foot with intrinsic muscle ischemia are the most common causes of traumatic pes cavus.

To distinguish anterior cavus from posterior cavus, measurement of the calcaneal pitch angle on the standing lateral roentgenogram is helpful.[15] This angle is measured from the horizontal to a line drawn along the plantar surface of the calcaneus (Fig. 1). A calcaneal pitch angle greater than 30° suggests calcaneocavus or posterior cavus, a condition characteristic of poliomyelitis patients who have cavus foot due to weak gastrocnemius and soleus muscles.[15] Calcaneocavus deformity is unlikely in CMT disease. Muscle wasting in CMT disease frequently occurs sequentially: first, the foot intrinsics; second, the peroneals; and, third, the anterior tibial muscle. The posterior tibial and the bulky gastrocnemius and soleus muscles are the last to atrophy.[14] Hindfoot equinus is most often seen in patients with CMT disease because of this unopposed action of the ankle plantar flexors. Thus, in contrast to patients with posterior cavus, in these patients the cavus deformity is secondary to muscular imbalances acting on the mid and anterior tarsus and the metatarsals. This is referred to as "anterior cavus," and on a lateral roentgenogram the calcaneal pitch angle is less than 30°.

PES CAVUS: MUSCULAR FACTORS

Muscles responsible for foot inversion and eversion contribute significantly to the cavus deformity in CMT. Hallgrimsson[6] and subsequently Mann and Missirian[12] postulated that overpull of the long peroneal results in plantar flexion of the first ray in these patients. This would account for the predominance of the first ray or forefoot valgus observed in many CMT patients when the hindfoot of the unloaded foot is held in the neutral position. In addition, Mann and Missirian[12] suggested that weakness of the short peroneal leaves the action of its antagonist, the posterior tibial muscle, unopposed, which further accentuates the high arch created by plantar flexion of the first ray.

The role of intrinsic muscle atrophy in the development of anterior cavus is not clear. Flexion deformities of the toes are frequently part of the foot deformity in CMT and can contribute significantly to the patient's complaints.[17] With intrinsic muscle atrophy,

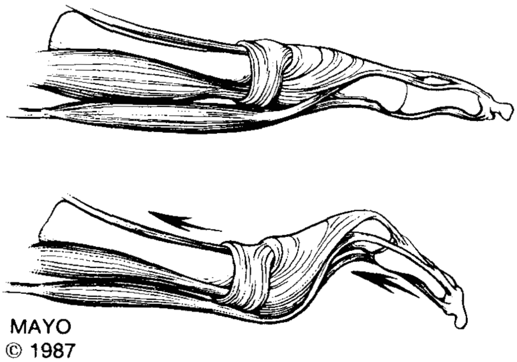


FIG. 2. Loss of intrinsic muscle action leads to extension deformity at the metatarsophalangeal joints and flexion at the interphalangeal joints. (Reproduced with permission from the Mayo Foundation.)

flexion at the metatarsophalangeal joints is lost, and the long toe extensors pull the toes into the claw position. The long toe flexors are able to flex only the interphalangeal joints, further accentuating the claw deformity (Fig. 2).

Prolonged muscle imbalance in CMT leads to bony deformity in skeletally immature patients; in adults, soft-tissue contractures result in rigid deformities. The toes become fixed, dorsally subluxated, and extended at the metatarsophalangeal joints and flexed at the interphalangeal joints. The forefoot assumes either a fixed valgus position (with the heel held in neutral) or all the metatarsals may be rigidly plantar-flexed, creating a deformity referred to as global metatarsus equinus.

FRONTAL PLANE DEFORMITIES

Forefoot deformities may have secondary effects on hindfoot alignment. Fixed plantar flexion of the first ray can result in hindfoot varus by what has been described as the tripod effect.[13] With forefoot valgus due to a rigidly plantar-flexed first metatarsal and mobile lateral rays, a plantigrade foot in midstance or standing depends on a varus roll of the heel to allow the lateral metatarsals to touch the floor (Fig. 3). Although flexible in the early stages, this hindfoot malalign-

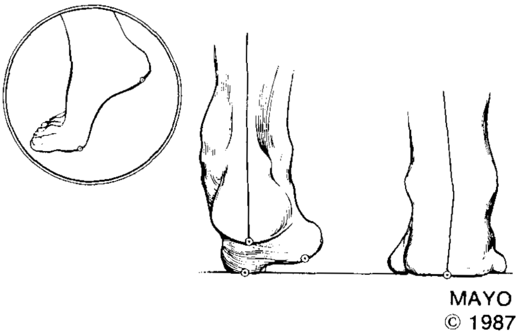


FIG. 3. Tripod effect. (Modified from Paulos *et al.*[13]; reproduced with permission from the Mayo Foundation.)

ment may eventually become fixed. The lateral block test devised by Coleman and Chesnut[3] is helpful in determining the flexibility of the hindfoot varus. This test is performed by placing a long block of wood under the heel and lateral rays, allowing the first metatarsal to drop to the floor (Figs. 4 and 5). Supporting the lateral metatarsals

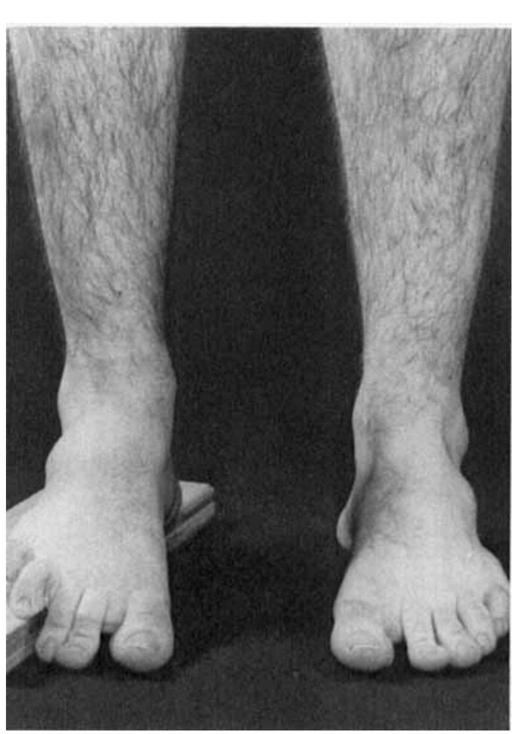
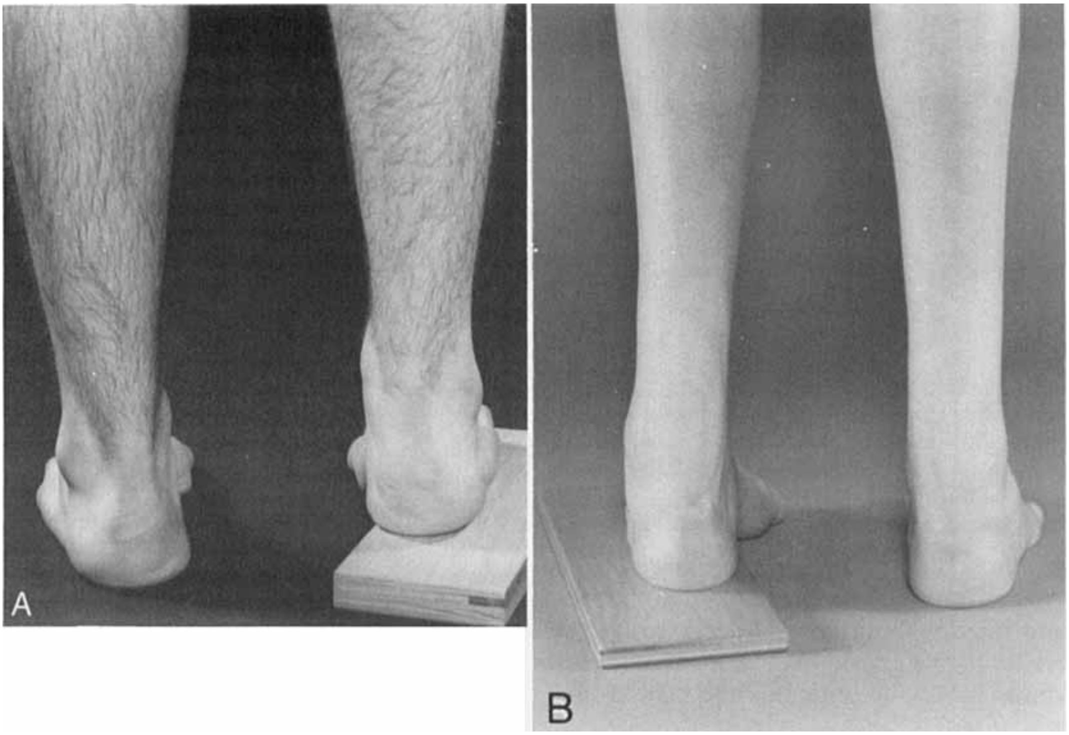


FIG. 4. Lateral block test, anterior view.



FIGS. 5A AND 5B. Lateral block test, posterior view. (A) Flexible hindfoot varus. (B) Fixed hindfoot varus.

eliminates the tripod effect and, if hindfoot varus is flexible, the heel will assume a neutral or valgus position. Persistent hindfoot varus with this test indicates that the hindfoot deformity is fixed.

CONSERVATIVE TREATMENT

Particularly in patients with mild involvement, pedorthic treatment can be effective. Initially, use of extra-depth shoes with support to unload the metatarsal heads is often helpful. Patients with a plantar-flexed first ray and compensatory flexible hindfoot varus may benefit considerably from use of a shoe or insert that posts the lateral forefoot and eliminates the inverting effect of forefoot valgus on the hindfoot. In advanced cases, molded shoes or lace-up custom-molded boots may be necessary. Some authors recommend a stretching program for flexible deformities, but its efficacy in this progressive neuromuscular condition is doubtful.

SURGICAL TREATMENT

Surgical management of the cavus foot resulting from the muscular imbalances of CMT disease must be individualized. There is no single surgical solution for the wide spectrum of involvement at presentation. Distinguishing between fixed and flexible components of the deformity is an essential step in the decision-making process. Tendon lengthening, transfer, and occasionally tenotomy, can be utilized to eliminate deforming forces and to balance the foot, but osteotomies and arthrodeses frequently are necessary to correct alignment and alleviate symptoms.

FOREFOOT

The most frequent forefoot deformity is clawing of the toes due to intrinsic weakness. Flexible or passively correctable claw toes often will resolve spontaneously with surgical correction of the midfoot cavus deformity.[4] It is advisable to observe the effect of midfoot cavus correction before treating the flexible toe deformity unless there is isolated toe involvement, as is occasionally seen in patients with CMT. Flexible claw toes without midfoot cavus and those that do not resolve spontaneously with midfoot correction are effectively treated by flexor-to-extensor transfers of the Girdlestone–Taylor type. Fixed lesser claw toes can be realigned by fusing the interphalangeal joints, transferring the long flexor dorsally, lengthening the extensors, and releasing the metatarsophalangeal joints. Fixed and flexible clawing of the great toe can be corrected reliably by the Jones procedure, which consists of an interphalangeal arthrodesis and long extensor transfer to the first metatarsal neck[7] (Fig. 6).

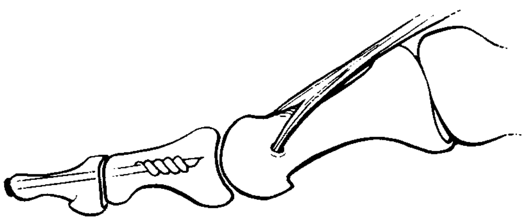


FIG. 6. Jones procedure for clawing of the great toe.

MIDFOOT

At the metatarsal level, the characteristic equinus deformity is either predominantly medial with plantar flexion of the first ray, creating a forefoot valgus deformity, or the plantar flexion involves the entire forefoot; the latter is referred to as global metatarsus equinus. In the early stages, patients with a progressive predominance of the first ray and somewhat supple forefoot valgus may benefit by removal of the plantar flexion force of the long peroneal on the first metatarsal. Transfer of the long peroneal to the short has yielded good results when preoperative rigidity was minimal.[5] In Hallgrimsson's[6] series of 51 cases, patient satisfaction improved with time after surgery, indicating that removing the deforming force may allow spontaneous stretching of the tight plantar tissues, particularly in younger patients. Rigid, long-standing plantar flexion of the first metatarsal with fixed forefoot valgus is best treated with a truncated closing wedge osteotomy of the first metatarsal through the proximal metaphysis (Fig. 7) or arthrodesis of the first tarsometatarsal joint with a wedge resection of the articular surfaces. Difficulty in opposing the osteotomy surfaces may necessitate release of the plantar fascia. Removing a truncated wedge may eliminate the need for plantar fascia release by considerably shortening the first metatarsal. Transfer of the long peroneal to the short as part of the procedure eliminates the deforming force and may improve hindfoot eversion.

Global forefoot equinus requires correction that traverses the foot. Corrective coronal plane osteotomies at multiple levels, from across the metatarsal bases to the transverse tarsal joint, have been described for this problem. Ideally, the most cosmetically acceptable result would be achieved by removing the corrective wedge at the apex of the cavus deformity. However, no publication to date has described a technique of determining the apex of the cavus or the implementa-

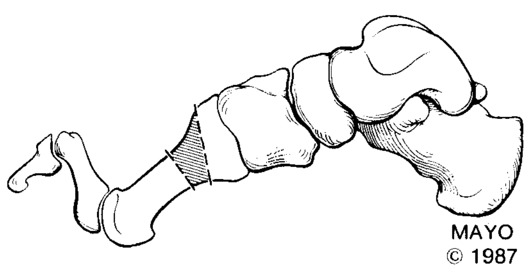


FIG. 7. Truncated wedge osteotomy of the first metatarsal. (Reproduced with permission of the Mayo Foundation.)

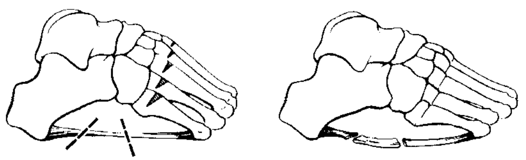


FIG. 8. Dorsally angulated greenstick osteotomy of the metatarsal bases.

tion of correction on the basis of this preoperative determination. Paper cutouts and, more recently, computer software packages designed to simulate corrective osteotomies may help in the preoperative planning process. However, these techniques are of limited value in the geometrically complex midfoot because they are two-dimensional. One can speculate that three-dimensional computed tomographic reconstructions will be the ultimate tool in planning these midfoot osteotomies.

Certain technical points are important to keep in mind in performing the osteotomies, and specific pitfalls must be avoided. As discussed previously in relation to the first metatarsal, simple closing wedge or greenstick osteotomies require a simultaneous plantar release to allow osteotomy closure. Removing a truncated wedge at any level decreases the need for plantar soft-tissue lengthening, but considerable shortening of the foot occurs. Overzealous wedge excision at any level of the midfoot can result in an overcorrected rocker-bottom foot.

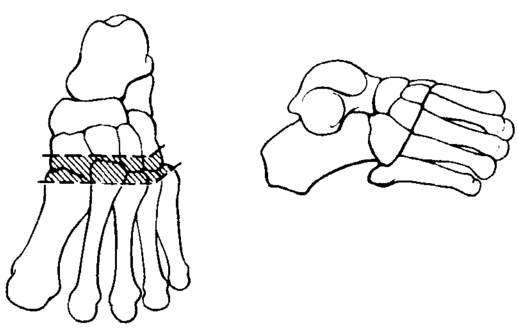


FIG. 9. Tarsometatarsal truncated wedge osteotomy. (Modified from Jahss.[8])

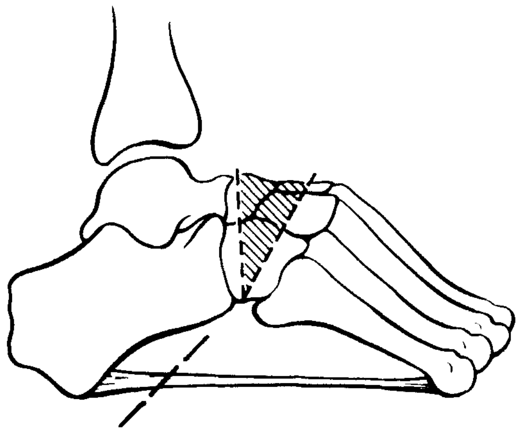


FIG. 10. The wedge osteotomy of Cole.[2]

The most distal of the bony procedures designed to correct forefoot equinus is a dorsally angulated greenstick osteotomy of the metatarsal bases (Fig. 8). This technique, introduced by Swanson *et al.*[17] and utilized by Gould,[5] has had reasonable success. A bayonet-shaped foot is more common after this procedure than after the more proximal osteotomies. The tarsometatarsal truncated wedge osteotomy was reviewed by Jahss,[8] who discouraged its use in CMT disease, stating a preference for triple arthrodesis in these patients (Fig. 9). The anterior tarsal wedge osteotomy (Fig. 10) popularized years ago by Cole[2] and, more recently, the V osteotomy of Japas[9] (Fig. 11) allow more proximal correction closer to the usual apex of the deformity. A pseudarthrosis rate of 30% reported[11] with the Cole osteotomy (Fig. 12) and marked widening of the midtarsal area with the V osteotomy[9] have resulted in many surgeons' avoiding cavus correction through the proximal midfoot region.

HINDFOOT

Hindfoot equinus secondary to the relative overpull of the gastrocnemius soleus complex is common in patients with CMT. In many cases, lengthening of the Achilles tendon will help achieve a plantigrade foot and

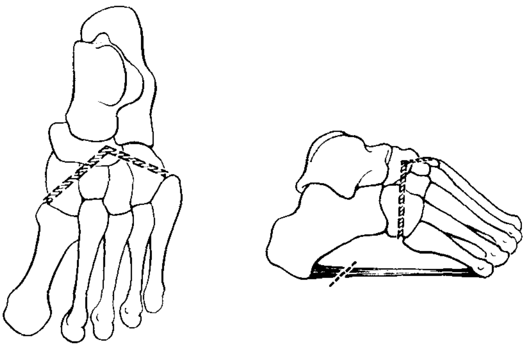


FIG. 11. The V osteotomy of Japas.[9]

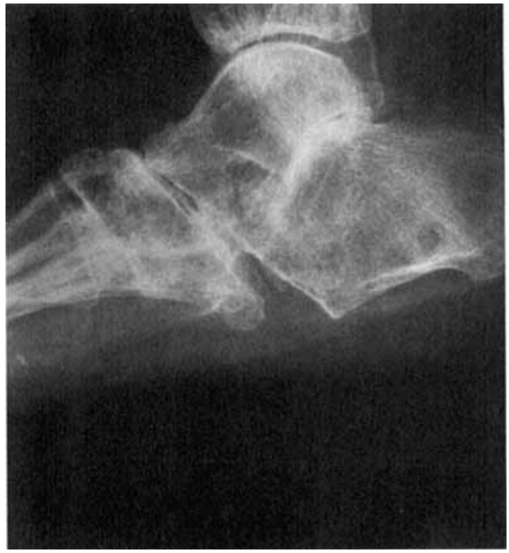


FIG. 12. A rocker-bottom foot with a nonunion of the tarsal wedge osteotomy.

decrease pressure on the metatarsal heads. Hindfoot varus is the malalignment usually associated with a cavus foot. The mechanism of this deformity and the use of the Coleman lateral block test to assess its flexibility have been discussed. Patients with hindfoot varus that corrects spontaneously on the lateral block test should have resolution of their hindfoot malalignment with surgical correction of fixed forefoot valgus.

If the hindfoot fails to correct on the lateral block test, a rigid deformity exists and realignment can only be accomplished by direct surgical approach to the deformity. Dwyer[4] popularized a closing lateral wedge osteotomy of the calcaneus to correct for hindfoot varus (Fig. 13). Combined simultaneous Dwyer osteotomy and greenstick osteotomies of the metatarsal bases for CMT patients with pes cavus has been described by Gould,[5] who achieved satisfactory results with this technique. The work of Levitt *et al.*[11] casts some doubt on the long-term effectiveness of combined calcaneal osteotomy and plantar fascial releases, as originally described by Dwyer, in the treatment of pes cavus due to progressive neurologic conditions.

Triple arthrodesis is a key procedure in the management of pes cavus due to CMT disease. The stage at which this stabilizing procedure should be instituted is controversial. In patients with severe rigid cavus deformity and marked limitation of subtalar mobility, triple arthrodesis represents the best surgical option. Correction of hindfoot varus is achieved through the subtalar (talocalcaneal) joint resection, and the cavus deformity is corrected through resection of the transverse tarsal joint. The technique of triple arthrode-

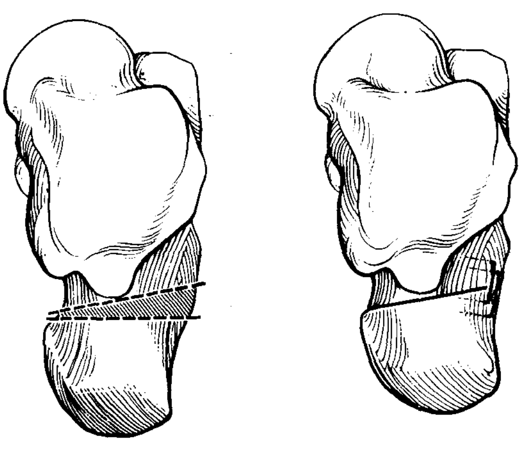


FIG. 13. Dwyer closing lateral wedge osteotomy of the calcaneus. (Modified from Gould.[5])

sis used depends primarily on the severity of the patient's deformity. For lesser degrees of equinus, conventional triple arthrodesis with corrective bone resection through the joints often will suffice. For advanced deformities the Lambrinudi triple arthrodesis or the beak arthrodesis will facilitate greater correction. Levitt *et al.*,[11] who demonstrated the effectiveness of triple arthrodesis as a salvage procedure when other procedures had failed to give permanent correction of pes cavus in patients with progressive neurologic disorders, were strong proponents of early triple arthrodesis in this subgroup of patients. They concluded that stabilizing the foot with a triple arthrodesis is essential to prevent progression of the deformity with further neurologic deterioration. Jahss[8] had a similar view, discouraging the use of the truncated wedge osteotomy of the tarsometatarsal joint in patients with CMT and advocating triple arthrodesis in this group instead.

DISCUSSION

Surgical intervention in the treatment of painful pes cavus in CMT disease requires meticulous planning. The inheritance patterns of CMT disease provide an opportunity to follow the development of the progressive cavus foot. Identifying members of affected families with progressive pes cavus in the early stages may facilitate performing muscle-balancing procedures before rigid deformity is established. In cases of rigid deformity, osteotomy with the necessary soft-tissue releases remains the mainstay of treatment. Recognizing first-ray-predominant equinus allows correction of the forefoot with less-extensive midfoot surgery. Various midfoot procedures exist for correction of global metatarsus equinus, but each has inherent pitfalls. Careful preoperative planning and accurate surgical technique are critical. The need for hindfoot correction can be determined by the lateral block test. In patients with fixed hindfoot varus, a closing lateral-wedge calcaneal osteotomy may be effective in balancing the foot.

Whether or not triple arthrodesis offers better long-term stability than combined midfoot and hindfoot osteotomies in the presence of progressive neurologic deterioration is controversial. Triple arthrodesis is the most appropriate procedure for patients with advanced rigid cavus deformity and no subtalar motion, especially with degenerative changes. Fixed claw toes frequently will require operative intervention, but flexible toe deformity often will correct spontaneously with operative reduction of the forefoot equinus.

Assessment of each component of the foot deformity and its flexibility is crucial in the effective management of pes cavus in CMT disease. Carefully planned surgical treatment usually will improve foot function and relieve symptoms secondary to the deformity. It is important, however, to make patients aware of the dynamic nature of their condition and the potential for deterioration of the foot correction with time.

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